

## Case reports

### Masson's pseudoangiosarcoma

G M Kavanagh MB MRCP<sup>1</sup> N Rooney MD MRCPath<sup>2</sup>  
C T C Kennedy MA FRCP<sup>1</sup> Departments of  
<sup>1</sup>Dermatology and <sup>2</sup>Pathology, Bristol Royal  
Infirmary, Bristol BS2 8HW

**Keywords:** benign pseudoangiosarcoma

Intravascular papillary endothelial hyperplasia (Masson's pseudoangiosarcoma) was first described in 1923<sup>1</sup> and received very little attention until 1974<sup>2</sup>. This lesion is distinguished by the presence of an exuberant endothelial proliferation within the distended lumina of medium sized veins. The pathogenesis of Masson's tumour remains speculative. At times it may simulate a low-grade angiosarcoma.

#### Case report

A 58-year-old man presented with an 8 month history of a painless, multinodular lesion over the right second MCP joint. There was a vague history of blunt trauma preceding the onset of this lesion. In the past, he had had a 'varicose vein' on the dorsum of the left hand since childhood which was excised in 1960. Histology was not obtained. Examination of the skin revealed a soft, lobulated, bluish swelling over the right second MCP joint. (see Figure 1).

A biopsy of this lesion revealed an abnormal vascular structure with a large central lumen containing thrombus in varying stages of organization. In several sections, particularly where thrombus formation was more developed, papillary fronds enveloped in not more than two layers of endothelial cell layers could be seen intimately associated with thrombus. These papillae fused in places to form characteristic slit-like spaces lined by endothelial cells with occasional hyperchromatic nuclei but no anaplasia. Immunostaining with von Willebrand factor and a new monoclonal antibody to endothelium, QBEND/10 (Quantum Biosystems,

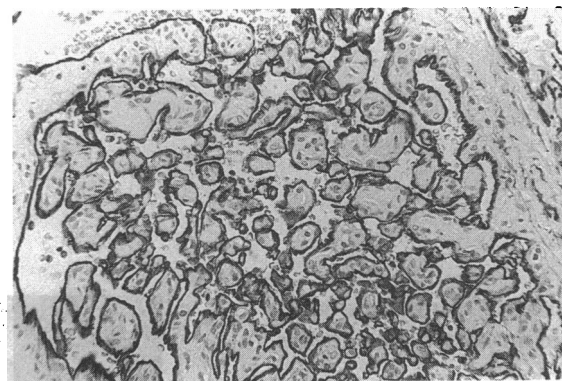


Figure 2. Papillary-like projections associated with a single layer of endothelial cells, stained with QBEND 10 antibody (H&E x75)

Cambridge, UK)<sup>3</sup>, confirmed these to be endothelial cells (see Figure 2).

#### Discussion

This benign vascular tumour, first described as a 'vegetating intravascular haemangioma'<sup>1</sup>, is characterized by several distinctive histological features which differentiate it from an angiosarcoma<sup>4</sup>. Confinement of the proliferative process to the intravascular space, lack of anaplasia and multiple mitoses, absence of true endothelial fronds and the presence of papillae consisting of fibrohyalinised tissue lined by not more than two endothelial cell layers, define this lesion. However, like angiosarcoma, it may show plump endothelial cells, papillary configuration, thrombus formation and irregular blood filled spaces.

These changes have been described in other body sites<sup>5</sup>. Masson's pseudoangiosarcoma typically occurs in medium sized veins but may also arise from pre-existing benign vascular tumours such as pyogenic granuloma, haemangioma and lymphangioma<sup>6</sup>. The lesion predominantly affects the head, neck and extremities and recurrence is unusual<sup>4</sup>. Angiolymphoid hyperplasia with eosinophilia and papular angioplasia have been included under the heading of 'pseudoangiosarcoma'<sup>7</sup> and share a predilection for the head, neck and extremities. However, there is wide variation among these conditions in both clinical and histological appearance.

In most instances, the lesion is found in association with a thrombus. Salyer *et al.*<sup>8</sup> compared the changes seen in Masson's pseudoangiosarcoma with those of angiosarcoma, and a large number of thrombi obtained at autopsy. All of the features described in Masson's tumour were observed in a small number of these thrombi. They concluded that this entity represented a peculiar form of organizing thrombus whereby endothelialization of thrombus fragments occurs to form papillary structures and slit like spaces. However, the occasional case described without thrombus, and the presence of similar changes in lymphatic vessels led Kuo *et al.*<sup>5</sup> to support Masson's original proposal that this was primarily endothelial proliferation with secondary thrombus formation.

This well defined entity is of interest to dermatologists, as it may present with non specific clinical appearances and shares certain histological features with angiosarcoma.

Case presented  
to Section of  
Dermatology,  
20 June 1991

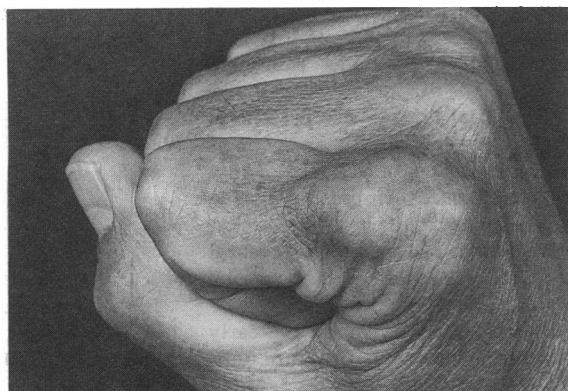


Figure 1. A soft, lobulated swelling over the right second MCP joint, which was bluish-purple in colour

0141-0768/91/  
120745-02/\$02.00/0  
© 1991  
The Royal  
Society of  
Medicine

## References

- 1 Masson P. Hemangioendothelioma vegetant intravasculaire. *Bull Soc Anat* 1923;23:517-23
- 2 Rosai J, Ackerman LR. Intravascular atypical vascular proliferation. *Arch Dermatol* 1974;109:714-17
- 3 Romani P, Bradley NJ, Fletcher CDM. QBEND/10, a new monoclonal antibody to endothelium: assessment of its diagnostic utility in paraffin sections. *Histopathology* 1990;17:237-42
- 4 Kuo TT, Sayers P, Rosai J. Masson's 'vegetant intra-vascular hemangioendothelioma': a lesion often mistaken for angiosarcoma. *Cancer* 1976;38:1227-36
- 5 Henschen F. L'endovascularite proliferante thrombopoietique dans la lesion vasculaire locale. *Ann Anat Pathol* 1932;9:113-21
- 6 Kuo TT, Gomez LG. Papillary endothelial proliferation in cystic lymphangiomas. *Arch Pathol Lab Med* 1979;103:306-8
- 7 Lever WF, Schaumburg-Lever G. *Histopathology of the skin*. 6th edn. Philadelphia: JB Lippencott, 1983:643-4
- 8 Salyer RW, Salyer DC. Intravascular angiomatosis: development and distinction from angiosarcoma. *Cancer* 1975;36:995-1001

(Accepted 3 July 1991)

## Infective endocarditis complicated by ruptured cerebral mycotic aneurysm

R L Patel FRCS<sup>1</sup> P Richards FRCS<sup>2</sup>D J Chambers PhD<sup>1</sup> G Venn FRCS<sup>1</sup><sup>1</sup>Department of Cardiothoracic Surgery, St Thomas' Hospital, London and <sup>2</sup>Department of Neurosurgery, Charing Cross Hospital, London**Keywords:** infective endocarditis; mycotic aneurysm; cerebral aneurysm; cerebral haemorrhage

A 31-year-old man with infective endocarditis of the aortic and mitral valves underwent double valve replacement, following which he initially made an uneventful recovery. Fifteen days following surgery he developed a rapidly progressive left hemiparesis due to an intra-cerebral bleed from a ruptured mycotic aneurysm. He underwent emergency craniotomy with evacuation of intracerebral haematoma and made a complete recovery, both from neurological and cardiological point of view.

### Case report

A 31-year-old man was admitted with a 4-month history of fever, night sweats, one stone weight loss, myalgia and arthritis. On examination he was pyrexial with a fever of 40°C, had splinter haemorrhages, finger clubbing, Janeway lesions, sub-conjunctival haemorrhages, splenomegaly and cardiac murmurs of aortic and mitral incompetence. A diagnosis of infective endocarditis was confirmed with the isolation of *Streptococcus viridans* in multiple blood cultures. Despite appropriate intravenous antibiotics, his fever persisted with progressive signs of cardiac decompensation necessitating emergency cardiac surgery. Both aortic and mitral valves were replaced with mechanical prostheses. He made an uneventful initial postoperative recovery with complete resolution of his pyrexia. Systemic antibiotic therapy continued in the postoperative period and anticoagulation was maintained within therapeutic range. On the twelfth postoperative day he developed recurrent fever, accompanied by rigors. Multiple blood cultures proved sterile and abruptly, on the third day of the return of fever, he developed a dense left hemiparesis, neck stiffness and left homonymous hemianopia.

CAT scan of the brain (Figure 1) revealed a large right posterior temporo-parietal haematoma. Right carotid angiogram demonstrated a distal middle cerebral mycotic

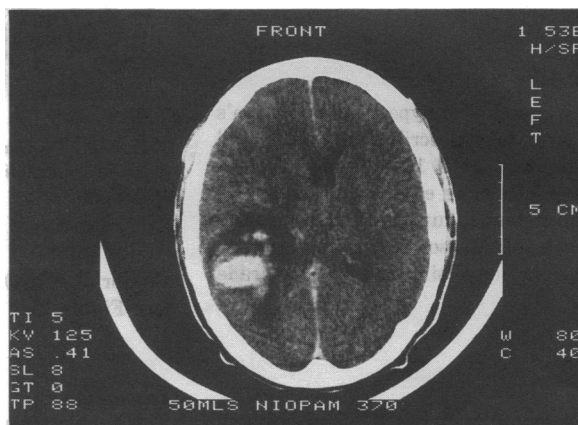


Figure 1. CAT scan of the brain showing large right posterior temporo-parietal intra-cerebral haematoma. There is surrounding oedema and shift of midline to the left

aneurysm as the source of the haematoma with two further small mycotic aneurysms related to the middle cerebral artery (Figure 2).

Despite full measures to reduce intracranial pressure, his right pupil began to dilate and an emergency craniotomy was performed. At operation a large, thick-walled, laminated haematoma was evacuated from the right parietal region. Culture of the haematoma proved sterile.

Following craniotomy he made a full neurological recovery. Six weeks after surgery four vessel cerebral angiography demonstrated complete resolution of the aneurysms demonstrated in the initial angiogram.

### Discussion

Neurological sequelae are recognized as some of the most life threatening complications of infective endocarditis in both treated and untreated patients. The incidence of neurological complications of between 20% and 40% in patients with infective endocarditis has remained remarkably constant.

Mycotic aneurysms of cerebral arteries are present in 2-10% of patients with infective endocarditis and account for 2.5-6.2% of all intracranial lesions of this type<sup>1</sup>. Those that rupture may destroy and obscure their presence during the massive haemorrhagic damage to the brain tissue that follows<sup>2-4</sup> and carries a fatality rate of 80%<sup>2</sup>. Cerebral embolization frequently precedes rupture of these mycotic aneurysms<sup>3,4</sup>.

Mycotic aneurysms often begin to develop in the early stages of infective endocarditis but may not rupture until many weeks or months after apparent recovery<sup>5,6</sup>. Many studies have documented, by serial angiography, the disappearance of cerebral mycotic aneurysms following successful antibiotic therapy<sup>4,7</sup>.

Case presented  
to Clinical  
Section  
12 April 1991